Sebaceous gland carcinoma usually arises from meibomian or Zeis glands deep within the eyelid, but it can rarely arise within the conjunctival epithelium without a deep component. We describe a woman with a history of chronic blepharoconjunctivitis unresponsive to topical medications. Examination disclosed confluent papillary hypertrophy of the upper palpebral conjunctiva and deposits of white flaky material. Tarsalconjunctival punch biopsy revealed intraepithelial sebaceous gland carcinoma. Management consisted of frozen section–controlled complete tumor excision with removal of the entire posterior lamella of the right upper eyelid, cryotherapy to the margins, and reconstruction. Histopathologic analysis confirmed primary sebaceous gland carcinoma localized to the conjunctival epithelium without involvement of underlying meibomian or Zeis glands or the caruncle. Patients with unexplained chronic unilateral blepharoconjunctivitis or papillary hypertrophy of the palpebral conjunctiva should be considered for biopsy to rule out neoplasia, even when there is no sign of an underlying eyelid mass.

Conjunctival intraepithelial sebaceous gland carcinoma typically arises from an underlying primary meibomian or Zeis gland carcinoma that secondarily invades the conjunctival epithelium by a centripetal or radial migration of tumor cells. This has been correlated with poor ocular and life prognosis. Three histopathologic patterns are exhibited by intraepithelial conjunctival sebaceous gland carcinoma—the bowenoid, pagetoid, and papillary types. The bowenoid type is characterized by full-thickness replacement of the epithelium by tumor cells that are large and pleomorphic and exhibit prominent mitotic activity. The pagetoid type is characterized by scattered individual tumor cells or nests of tumor cells within the epithelium that are devoid of intercellular bridges. The papillary pattern is rare and manifests with intraepithelial confluent cells resembling carcinoma in situ.

In 1967, Theodore and Irvine described a “masquerade syndrome” characterized by chronic blepharoconjunctivitis due to an unsuspected conjunctival intraepithelial squamous cell or sebaceous gland carcinoma. Although the originally described neoplasms were squamous cell carcinomas, in retrospect, many of the tumors producing this clinical picture may have been sebaceous gland carcinomas. Others have also emphasized that tarsalconjunctival inflammation is common in patients with intraepithelial sebaceous gland carcinoma of the conjunctiva.
involving the inferior tarsal and bulbar conjunctiva, with one small focus of invasive tumor of the bulbar conjunctiva. There was extensive scarring of the tarsus but the meibomian and Zeis glands showed no clear-cut source of tumor. Margo and Grossniklaus\(^\text{10}\) later reported 2 similar cases in a 58-year-old woman and a 71-year-old man, both treated with orbital exenteration. The meibomian and Zeis glands showed no carcinoma but were completely replaced by lipogranulomatous inflammation. Our case was unique in several respects. The tumor occurred in a young patient aged 33 years who had been receiving oral corticosteroids. Immunosuppression may have contributed to the young age at onset of sebaceous neoplasia, as the patient had no other known predisposition, such as radiation exposure.\(^\text{16}\) In addition, there was no histopathologic evidence of inflammation, scarring, or tumor in the underlying sebaceous glands. The lack of underlying tarsal tumor on histopathologic examination is extremely unusual and raises speculation as to the source of the malignant cells.

The origin of primary conjunctival intraepithelial sebaceous gland carcinoma without deep involvement has been debated.\(^\text{8-10}\) One argument is that the conjunctival epithelium has the potential to spawn sebaceous gland carcinoma.\(^\text{8}\) From an embryologic point of view, this is understandable, as the sebaceous glands of the tarsus and caruncle arise from invaginations of the embryonic conjunctival epithelium.\(^\text{17}\) To support this hypothesis, 2 cases of papillomas of the tarsal conjunctival epithelium with focal sebaceous differentiation have been identified.\(^\text{8}\) Another theory suggests that glandular sebaceous neoplasms could give rise to intraepithelial spread on the ocular surface followed by spontaneous involution of the glandular component, leaving only intraepithelial disease.\(^\text{10}\) Last, it should be realized that the presence of a focus of microinvasive or deep glandular tumor cannot fully be eliminated, even by step sectioning of the specimen, as the tumor is known to have skip areas.

The optimal method of treating intraepithelial sebaceous gland carcinoma is controversial. Suggested modalities include careful observation, cryotherapy, radiotherapy, complete excision, and orbital exenteration.\(^\text{8-10}\) We chose complete excision with frozen section control and cryotherapy after map biopsies disclosed no tumor in the remainder of the conjunctiva.

Figure 2. A, The excised tarsoconjunctival lamina showing full-thickness replacement of conjunctival epithelium by sebaceous gland carcinoma. Note the absence of deep focus in the meibomian glands or an invasive component (hematoxylin-eosin, original magnification \(\times20\)). B, Large atypical cells with vacuolated cytoplasm and large vesicular nuclei, diagnostic of sebaceous gland carcinoma (hematoxylin-eosin, original magnification \(\times200\)). C, Frozen section of the palpebral conjunctiva showing positive oil-red-O staining indicating the presence of intracellular lipid (oil-red-O, original magnification \(\times200\)).

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In conclusion, we describe a patient with primary sebaceous gland carcinoma of the conjunctival epithelium without evidence of involvement of the tarsus, Zeis glands, caruncle, or other sites of normal sebaceous glands. Careful follow-up is necessary as the primary tumor site may not yet be evident. Any patient with unexplained asymmetric, chronic blepharconjunctivitis or papillary hypertrophy of the palpebral conjunctiva should be considered for biopsy to rule out sebaceous gland carcinoma, even in a young patient.

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REFERENCES


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A look at the past...

...when Col Smith introduced his operation for the intracapsular extraction of cataract, ophthalmic surgeons were much struck by the beauty of the operation, and the skill of the originator. Sir John Parsons referred to his visit to St Louis and watching Dr Green operate by the Col Smith method. He was not favorably impressed by the results; at the same time he thought “show operations” were a great mistake, as, under these conditions, it was difficult to fairly assess the value of a particular operation. His own view was that intracapsular methods must of necessity be accompanied by increased dangers to the patient’s eye.